250 Bronchodilator responsiveness in infants with cystic fibrosis in Argentina

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Introduction: The prevalence of bronchodilator (BD) responsiveness in children with Cystic Fibrosis (CF) is variable; inclusive during the longitudinal follow-up of the same patient.
Objective: To assess bronchodilator responsiveness in infants with CF younger than 24 months assisted at the Center of Cystic Fibrosis in La Plata.
Materials and Methods: Cross sectional study. We measured: Functional Residual Capacity (FRC), Airway Resistance (Raw) and specific conductance (sGaw) by plethismography, and Maximal Expiratory Flow at FRC (V′maxFRC) using rapid thoracic compression with Jaeger® equipment, before and after 15 minutes of the inhalation of 200 mcg of salbutamol (Pre BD and Post BD, respectively). It was considered to be significant bronchodilator response to changing V'maxFRC of 30% over the prebroncodilator value. Paired t-test was performed to compare the means of the indicators using the SPSS software 9.0.
Results: We evaluated 20 patients with a mean age of 15.9 months. Fourteen completed the assessment of the post BD. Eight (57%) showed an increase and six (43%) a decrease in flows. Four patients (28.5%) had a significant response to bronchodilator. The FRC and Raw decreased and sGaw increased after BD, but without significant differences.
Conclusions: These results suggest that the evaluation of the response to bronchodilators should be considered in the treatment of infants with CF. Moreover, possible changes in a longitudinal long term follow-up should also be evaluated.

	V′maxFRC (ml/s)	Z score V′maxFRC	FRC (ml)	Raw	sGaw
n	14	14	11	12	12
Pre BD	186	−1.05	246	1.72	1.10
Post BD	187	−0.99	240	1.03	1.94

251 Spirometry in CF patients

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Introduction: Maintenance of pulmonary function is a key aim in the treatment of CF patients, and as part of this an accurate measurement of spirometry is crucial when assessing the condition. In turn, spirometric measurement is only accurate if the forced manoeuvre technique is correct, multiple reproducible traces are performed and the effort is maximal, and in the UK guidelines indicates that it should be carried out by trained and accredited pulmonary physiologists, using calibrated equipment. We therefore reviewed the spirometric practice of CF units in the UK, looking at who performed the tests and where they were carried out.
Method: A telephone survey of 32 (of 49) CF clinics (20 adult) listed by the UK CF Trust as large CF centres (>50 patients).
Results: Three units did not involve pulmonary physiologists: 2 adult (both physiotherapist led) and 1 paediatric (nurse led).
In the remaining 29 units, for outpatient spirometry: pulmonary physiologists alone (15), physiotherapists alone (2), nurses alone (3), the remainder a mixture of all 3 groups; in the pulmonary function department (15), outpatient clinic (9), the remainder a mixture.
For inpatients: pulmonary physiologists alone (10), physiotherapists alone (1), nurses alone (5), in the remainder a combination of personnel; on the ward alone (7), pulmonary function department alone (11), in both locations for the remainder.
Conclusions: This audit shows that some CF units in the UK do not involve pulmonary physiologists in the measurement of spirometry, and many other units allow tests to be carried out by personnel who are unlikely to be trained to an accredited standard, calling into the question the accuracy of the measurements produced. A national review of spirometric practise may help to correct these deficiencies.

252 Correlation between pulmonary function and nutritional status, age, and bacterial colonization in cystic fibrosis (CF) patients

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CF is the most frequent genetic disorder in Caucasian population. Pulmonary function, age, bacterial colonization and nutritional status are main factors related with prognosis in CF patients. The aim of the study was to correlate the declination of the FEV1 with nutritional status, age and bacterial colonization in CF children.
Materials and Methods: observational and cross-sectional study. We invited to participate all patients with CF over 5 years old capable to perform a pulmonary function test. Forty three patients were included. They were divided in four groups according of their baseline FEV1. Age, Z-score body mass index (BMI), and bacterial colonization (three or more sputum cultures positive to any bacteria) were the variables analyzed.
Results: See the table. The FEV1 showed a positive correlation with Z-score BMI ($r = 0.7$; $p < 0.01$) and an inverse correlation with age ($r = -0.56$; $p < 0.01$). The Mantel Test showed a significant tendency of proportions between the normal pulmonary function group and the others to colonization with methicillin resistant *Staphylococcus aureus* ($p < 0.01$), *Burkholderia cepacia* ($p = 0.02$), and *Pseudomonas aeruginosa* ($p = 0.05$).
Conclusions: pulmonary function declines with age, bacterial colonization and malnutrition in CF children.

Groups	⩾80%	60–79%	40–59%	⩽39%
n (m/f)	17 (10/7)	9 (6/3)	9 (3/6)	8 (5/3)
age (years)	8.5±3	10.9±4	12.2±5	14.0±4
FEV1 (%)	103±15	69±7	50±7	31±7
Z-Score BMI (range)	0.23 (−1.4–1.5)	−0.38 (−1.39–0.65)	−0.89 (−3.39–0.32)	−1.76 (−3.15–0.46)
Bacterial colonization	71%	77%	100%	100%
P. aeruginosa	41%	55%	77%	75%
Met R *S. aureus*	6%	45%	56%	88%
B. cepacia	6%	11%	33%	38%

253 Correlation between respiratory function tests, scintigraphy and high-resolution computed tomography findings in patients with cystic fibrosis

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Objective: The aim of this prospective study is to find out the correlation between respiratory function tests (RFTs), scintigraphy (S) and high resolution computed tomography (HRCT) findings in patients with cystic fibrosis (CF).
Method: 17 patients (mean age of 12±2 years) with a diagnosis of CF were included. All patients had RFTs, ventilation (V)–perfusion S and HRCT. Scintigraphic images were scored based on defect severity and extent. HRCT images were scored for the degree of bronchiectasis (B), peribronchial thickening (BT), mucus plugging (MP), air trapping (AT), bullae and atelectasis/consolidation (A/C).
Results: There were 9 patients with normal perfusion and 7 patients with normal ventilation S and 8 patients had both abnormal ventilation and perfusion S. There were no significant difference in the ventilation and perfusion scores of patients (Mann Whitney U p: 0.980). The perfusion and ventilation S findings showed good correlation with HRCT. There was a significant correlation between perfusion scores and total HRCT, B, PT, MP, AT and A/C scores (0.839, 0.884, 0.803, 0.59, 0.508 respectively (Spearman correlation coefficient [SCC]).
There were no significant correlations between the imaging scores and RFTs (FEV1, raw and FEF25–75%). The only significant correlation was found with RV/TLC with SCCs of 0.645, 0.673, 0.715, 0.679, 0.613, 0.715, 0.595 for perfusion, ventilation, total HRCT, B, PT, MP, AT scores respectively.
Conclusion: The scintigraphic and HRCT findings showed a good correlation with each other and RFT parameter, RV/TLC in patients with CF.